Anesthetic management of a parturient with Ehlers–Danlos syndrome posted for elective cesarean section

Della Vella Beatrice, Luca Titi, Maria Grazia Tarsitano, Maria Luisa Borgia

Department of Anesthesiology, Critical Care and Pain Medicine, Section of Gynecology and Obstetrics, Sapienza University of Rome, Rome, Italy

Address for correspondence:
Dr. Luca Titi,
Department of Anesthesiology, Critical Care and Pain Medicine, Section of Gynecology and Obstetrics, Sapienza University of Rome, Rome, Italy
Via Spinetti Snc - 03021, Amaseno (FR), Italy.
E-mail: lucatiti@hotmail.it

ABSTRACT

A heterogeneous group of inherited disorders of the connective tissue, characterized by skin hyperextensibility, joint hypermobility and tissue fragility, is defined as Ehlers–Danlos syndrome (EDS). The clinical manifestations vary depending on the type of disease. This syndrome may be associated with numerous pregnancy complications, ranging from mild articular discomfort to maternal death. We report the anesthetic management and the literature review of a parturient with EDS undergoing elective cesarean section.

Key words: *Cesarean section, Ehlers–Danlos syndrome, general anesthesia, spinal anesthesia*

INTRODUCTION

A heterogeneous group of connective tissue diseases (transmitted genetically) caused by a deficiency in the synthesis of collagen, characterized by varying degrees of hyperextensibility skin, joint hypermobility and vascular fragility, musculoskeletal pain, joint dislocations, easy bleeding, atrophic scars, vessel/viscera rupture, severe scoliosis and obstetric complications, is called Ehlers–Danlos syndrome (EDS).[1] The features of this connective tissue disease with genetic transmission that have the most impact on the obstetric and anesthetic management of these pregnant patients include fragility, poorly healing skin, excessive bleeding, spontaneous pneumothorax, easy joint dislocation, valvular prolapse and spontaneous vascular dissections or ruptures of major vessels.[2] In the literature, there is little knowledge about the management of anesthesia during elective cesarean section in patients with EDS. We report our experience about elective cesarean section in a primigravid patient with EDS.

Access this article online	
Quick Response Code:	Website: www.joacc.com
	DOI: 10.4103/2249-4472.132822

CASE REPORT

A 35-year-old woman with EDS (weight 66 kg, height 1.66 cm) with full-term pregnancy and in ASA category III was posted for an elective cesarean section. She had no history of diabetes, hypertension or any other medical disorder. All relating coagulation studies (prothrombin time, activated partial thromboplastin time to fibrinogen and factor assays) and laboratory values (hemoglobin, hematocrit and platelet count) were normal. Physical examination of her airway revealed a Mallampati class III with adequate thyroid–chin distance and full range of motion of the head and neck.

The patient at the age of 15 years was investigated for low back pain, anterior knee pain and multiple subluxation/dislocation of both the hips. At that time, joint hypermobility and skin hyperextensilibity were also present. An analysis of the α2-chains of type I collagen gene (COL1A2) showed a mutation donor splice site. Based on these findings, a diagnosis of EDS was made. There was however no family history of

such a disease. After a prolonged detailed discussion, with both the patient and her husband, concerning problems that could occur as a result of regional anesthesia in a patient with bleeding problems, cervical spine instability and possibility of airway trauma during intubation, it was decided to proceed with spinal anesthesia. Preloading (20 mL/kg) was carried out using Ringer's lactate solution; under full aseptic precautions, spinal anesthesia was administered with 6.5 mg hyperbaric bupivacaine, sufentanil 2.5 mcg and 50 mcg morphine at the lumbar 4-5 interspace using a 25-Gauge spinal needle. While positioning (left lateral) for spinal anesthesia, flexion of the neck was avoided. The patient was then placed with a 15 degree tilt and oxygen (5l/min) was administered via a face mask. A small pillow was placed under the patient's head and left lateral tilt was maintained. Surgery was commenced only after the achievement of a sensory block till the level of T6 dermatome. The surgery lasted 50 min. The tital blood loss was about 350 mL and a female baby was extracted weighing 2.9 kg and with Apgar scores of 8 and 9 at 1 and 5 mins after birth.

DISCUSSION

EDS is a heterogeneous group of inherited connective tissue disorders caused by a defect in the synthesis of collagen, characterized by varying degrees of skin hyperextensibility, joint hypermobility and vascular fragility. Musculoskeletal pain, joint dislocations, easy bleeding, atrophic scars, vessel/viscera rupture, severe scoliosis and obstetric complications may occur.[1]

There is little information regarding preferred mode of delivery for these parturients. Cesarean section in these patients can be associated with an increased risk of perioperative hemorrage (due to uncontrolled bleeding from friable vessels and coagulopathy), impaired wound healing and a likelihood of wound dehiscence.[2]

Patients with this syndrome can also have aortic regurgitation, mitral valve prolapse and conduction disturbances (atrial fibrillation secondary to mitral regurgitation and atrial enlargement), sinus bradycardia and grade 1 atrioventrucular block. Invasive cardiac monitoring is likely to be essential in such cases.[3,4] Our patient did not have any of these cardiac lesions; therefore, invasive monitoring was not instituted.

The choice of anesthesia for a cesarean section is controversial as both general anesthesia and regional anesthesia can be given, and each of these techniques can be associated with complications. General anesthesia may be safer in view of the rapid onset and avoiding of bleeding after neuraxial block (spinal/epidural hematoma). It is well established that the maternal mortality rate is 16-times higher in pregnant patients who undergo abdominal delivery under general anesthesia as compared with those who receive regional anesthesia.[1,2,5-7] Managing airway while administering a general anesthetic in patients with EDS would be all the more difficult due to the presence of cervical spine joint laxity, unstable cervical spine (that would make laryngoscopy and intubation hazardous),[5,6] risk of cervical atlantoaxial subluxation and temporomandibular dislocation[2] and fragility of the oral and laryngeal tissues. Intraoperatively, low airway inflation pressures should be used because of the increased risk of pneumothorax.[5] Spontaneous ventilation seems attractive but has never been practiced.

In case a neuraxial blockade is chosen, both spinal and epidural blocks can be given. However, the presence of kyphoscoliosis and the increased elasticity of the spinal ligaments can make the administration of such blocks difficult (increased elasticity of the ligaments around the spine can make it harder for the anesthesiologist to feel the needle advancement).[8] Neuraxial techniques using epidural anesthesia and combined spinal–epidural anesthesia may be associated with an increased risk of epidural hematoma formation because of catheter insertion in a patient with increased fragility of the epidural veins. Furthermore, neuraxial block has been reported to have a reduced effect of anesthetics on patients with EDS and, therefore, it is recommended to combine several different types of anesthetic drugs (local anesthetic and opioid drugs) in order to achieve a sufficient anesthetic effect.[9]

There have been reports of resistance to local anesthetic agents in women with EDS.[10] Although the pathophysiology of this phenomenon remains unresolved, it is important to know this condition. Nonsteroidal drugs are best avoided in such patients due to the risk of gastric bleeding and, ideally, the postoperative pain should be treated with a combination of opioids and acetamophen.[11] Our patient was administered a single shot of spinal block that was uneventful.

CONCLUSIONS

Results of the study demonstrate that spinal anesthesia can be safely adminsitered for cesarean section in patients with EDS.

REFERENCES

1. Shirley ED, Demaio M, Bodurtha J. Ehlers-danlos syndrome in orthopaedics: Etiology, diagnosis, and treatment implications. Sports Health 2012;4:394-403.
2. Volkov N, Nisenblat V, Ohel G, Gonen R. Ehlers-Danlos syndrome: Insights on obstetric aspects. Obstet Gynecol Surv 2007;62:51-7.
3. Dill-Russell P, Jones LS. Anaesthesia for caesarean section in a patient with Ehlers-Danlos syndrome and mitral valve prolapse. Int J Obstet Anesth 2001;10:192-7.

4. Jones TL, Ng C. Anaesthesia for caesarean section in a patient with Ehlers-Danlos syndrome associated with postural orthostatic tachycardia syndrome. Int J Obstet Anesth 2008;17:365-9.
5. Lane D. Anaesthetic implications of vascular type Ehlers-Danlos syndrome. Anaesth Intensive Care 2006;34:501-5.
6. Halko GJ, Cobb R, Abeles M. Patients with type IV Ehlers-Danlos syndrome may be predisposed to atlantoaxial subluxation. J Rheumatol 1995;22:2152-5.
7. Dutta I, Wilson H, Oteri O. Pregnancy and delivery in ehlers-danlos syndrome (hypermobility type): Review of the literature. Obstet Gynecol Int 2011;2011:306413.
8. Randalls B, Broadway JW, Browne DA, Morgan BM. Comparison of four subarachnoid solutions in a needle-through-needle technique for elective caesarean section. Br J Anaesth 1991;66:314-8.
9. Arendt-Nielsen L, Kaalund S, Bjerring P, Høgsaa B. Insufficient effect of local analgesics in Ehlers Danlos type III patients (connective tissue disorder). Acta Anaesthesiol Scand 1990;34:358-61.
10. Sood V, Robinson DA, Suri I. Difficult intubation during rapid sequence induction in a parturient with Ehlers-Danlos syndrome, hypermobility type. Int J Obstet Anesth 2009;18:408-12.
11. Rombaut L, Malfait F, De Wandele I, Cools A, Thijs Y, De Paepe A, *et al*. Medication, surgery, and physiotherapy among patients with the hypermobility type of Ehlers-Danlos syndrome. Arch Phys Med Rehabil 2011;92:1106-12.

Cite this article as: Beatrice DV, Titi L, Tarsitano MG, Borgia ML. Anesthetic management of a parturient with Ehlers-Danlos syndrome posted for elective cesarean section. J Obstet Anaesth Crit Care 2014;4:35-7.

Source of Support: Nil, **Conflict of Interest:** None declared.